Congenital Heart Disease

Comments Regarding Incidence and Natural History

FORREST H. ADAMS, M.D., Los Angeles

• **The incidence of congenital heart disease is approximately 1 per cent of all live births. Approximately 60 per cent of patients who die of congenital heart disease do so at less than two years of age. Very few patients with such lesions live beyond 45 years at the very most. In about 70 per cent of patients who are born with cardiac anomalies, the lesions are either of kinds that are already being operated upon successfully or for which operations are now being attempted and often are helpful.**

IMPROVEMENTS IN THE diagnosis and treatment of patients with congenital heart disease have come swiftly. Even investigators working full time in the field find it difficult to keep up with the frequent advances. Oddly, however, little is known about prevention and until very recently not much was known about the incidence or the natural history of the various forms of congenital heart disease. Even now this information is fragmentary and hardly permits more than a few general statements.

Estimates of the frequency of congenital heart disease which are generally quoted have been based either upon clinical signs elicited during physical examination of older children, or upon autopsy data, a source which makes hospital deaths from all causes the only basis for comparisons. Richards and co-workers,[10] at Babies Hospital, Columbia University, studied the frequency of congenital heart disease in an unselected newborn population produced from 5,964 consecutive pregnancies. Table 1 shows the incidence of congenital heart disease for the 6,053 infants, grouped according to weight at birth. Fifty of them were born with congenital heart disease, an incidence of 0.83 per cent. This proportion may be slightly low since the living infants were observed for only one year; and it was pointed out that in some cases heart disease probably was "overlooked because of failure to develop symptoms or signs before the end of the follow-up." Richards said that it is reasonable to suppose that atrial septal defects may comprise the major portion of the lesions unrecognized under one year of age.

TABLE 1.—*Incidence of Congenital Heart Disease (CHD) in 6,053 Newborns (Richards[10])*

Weight of Newborn	Total	With CHD	
		No.	Per Cent
500 gm. or less	314	1	0.3
500 to 2,500 gm.	597	18	3.0
2,500 or more	5,142	31	0.6
Total	6,053	50	0.8
Surviving 1 month	5,530	38	0.6

TABLE 2.—*Survival of Patients with Congenital Heart Disease (Abbott[1])*

Condition	No. of Cases	Age		
		Maximum	Minimum	Mean
Atrial defect:				
Above	10	64 yr.	6 mo.	34 yr.
Below	18	46 yr.	8 mo.	19 yr.
Patent ductus arteriosus	92	66 yr.	2 wk.	24 yr.
Ventricular defect	50	49 yr.	Fetus	14 yr.
Coarctation of aorta:				
Adult	70	92 yr.	3 yr.	33 yr.
Infantile	9	9 mo.	8 hr.	2 mo.
Pulmonary stenosis	16	57 yr.	4 yr.	18 yr.
Tetralogy of Fallot	85	60 yr.	11 days	12 yr.

With this information as a base regarding the incidence, what is the natural history of patients with congenital heart disease? When do they become ill, and when do they die? The classic work in this regard is that of Abbott.[1] For no practical reason at the time, she set about collecting data on cases in which autopsy was done. In an excellent monograph she summarized the material she had gathered on the first 1,000 cases. Survival data on the most common congenital cardiac anomalies (as abstracted from her monograph) are given in Table 2.

It is known from at least two sources[6,9] that 60 per cent to 80 per cent of patients dying with congenital heart disease do so during the first two years of life. Recently, Boesen[4] of Denmark collected data on 1,145 infants and children under four years of age who died of congenital heart disease in all of Scandinavia. The original material has been rearranged by me into three groups: Those in which the lesions were operable, those in which operation was a possibility, and those in which operation cannot be done at present. As shown in Table 3, about a third of the patients had lesions of a kind for which corrective or palliative operation is of proved value.

From the Department of Pediatrics, University of California, Los Angeles.

Submitted July 25, 1958.

TABLE 3.—*Cause of Death Under 4 Years of Age (1,145 Cases);* Operable Lesions*

Diagnosis	Cases	Age at Death Less Than			
		30 Days	150 Days	1 Year	4 Years
Coarctation of aorta	83	40	70	80	3
Patent ductus arteriosus	70	24	54	65	5
Tetralogy of Fallot	48	15	24	36	12
Atrial defect (secundum)	35	10	26	31	4
Atrial defect (primum)	13	1	7	11	2
Aortic stenosis, valvular	30	14	25	29	1
Tricuspid stenosis	30	11	21	28	2
Atrial defect + patent ductus arteriosus	28	19	26	27	1
Pulmonary stenosis	19	9	11	16	3
Total	356	143	264	323	33

*Adapted from Boesen and Vendel: VIII International Pediatric Congress.

TABLE 4.—*Cause of Death Under 4 Years of Age (1,145 Cases);* Operation Possible*

Diagnosis	Cases	Age at Death Less Than			
		30 Days	150 Days	1 Year	4 Years
Ventricular defect	148	51	107	142	6
Ventricular defect + patent ductus arteriosus	65	34	54	63	2
Ventricular defect + atrial septal defect	15	1	10	14	1
Ventricular defect + patent ductus arteriosus + atrial septal defect	12	5	10	12	
Pulmonary stenosis + atrial septal defect/ventricular septal defect/patent ductus arteriosus	33	13	24	29	4
Atrioventricular communis	33	6	22	33	
Complete transposition	167	78	138	163	4
Single ventricle	90	52	80	88	2
Total	563	240	445	544	19

*Adapted from Boesen and Vendel: VIII International Pediatric Congress.

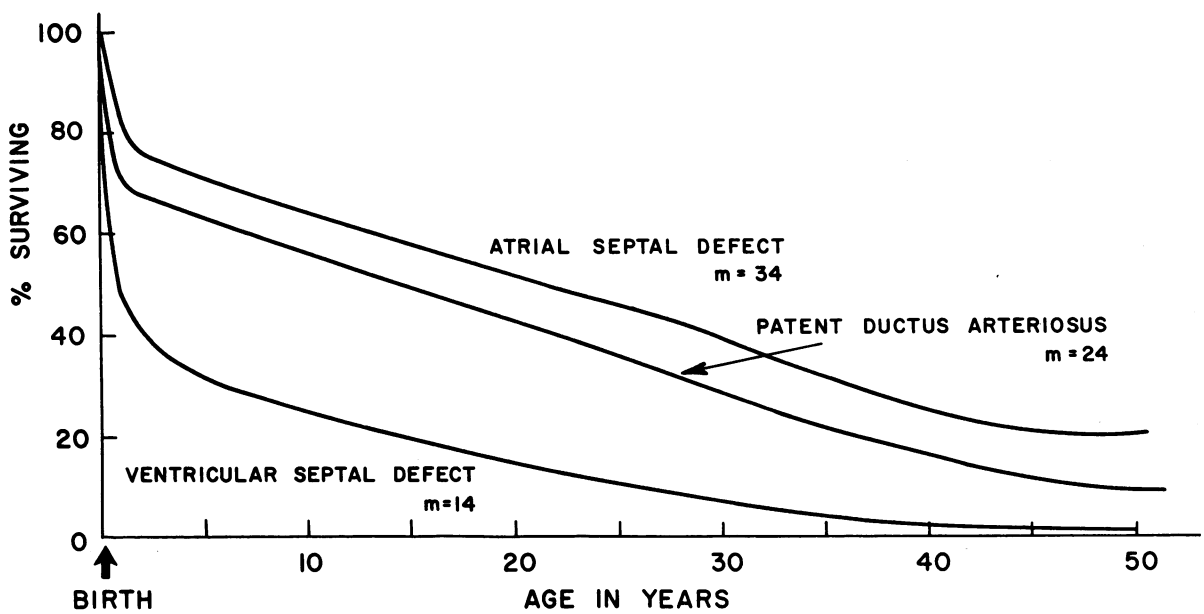

Chart 1.—Theoretical survival curves for patients with left to right shunt (mean age (m) obtained from Abbott[1]).

About half of the 1,145 patients were in the possibly operable group—that is, they had lesions for which operation is now being attempted and is either curative or palliative in many instances.

In order to visualize the natural history of some of these congenital heart lesions, I have made two diagrams which are purely theoretical but make use of the best information currently available. Chart 1 shows the theoretical survival curves for patients with left to right shunts and Chart 2 shows the curves for patients with stenotic lesions.

Congenital heart disease is not only significant in the cause of death in infancy but it is also a significant cause of morbidity. Keith[7] recently reviewed his experience in the diagnosis and treatment of heart failure in the pediatric age group. He reported on 304 patients, of whom 90 per cent were under one year of age. Chart 3 shows the causes of heart

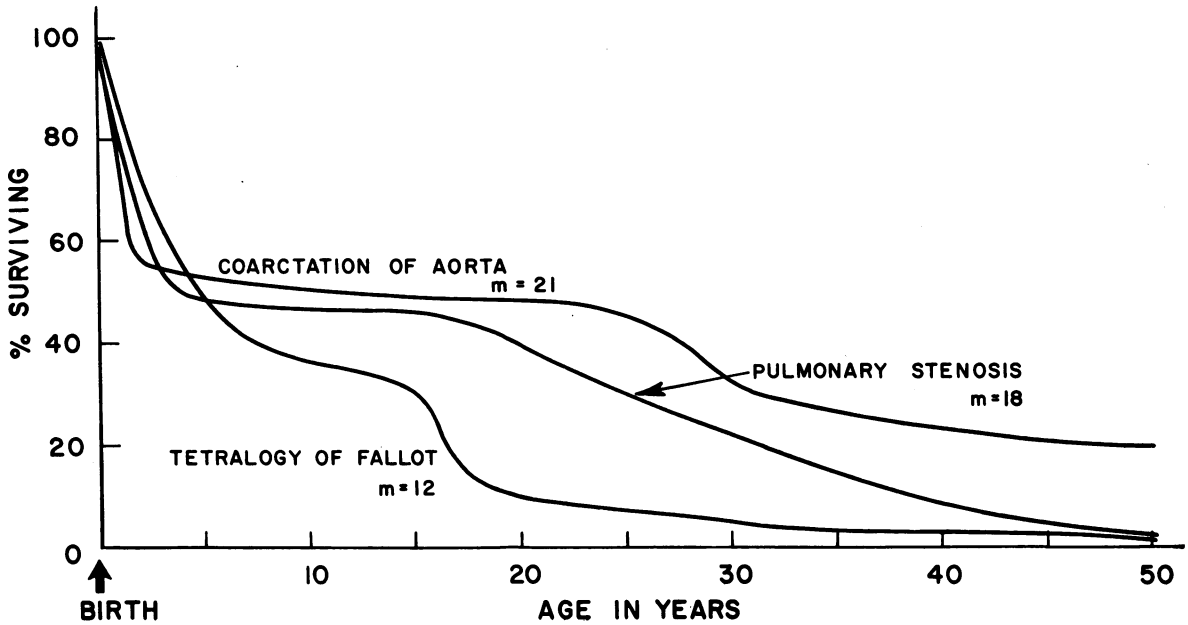

Chart 2.—Theoretical survival curve for patients with stenosis (mean age (m) obtained from Abbott[1]).

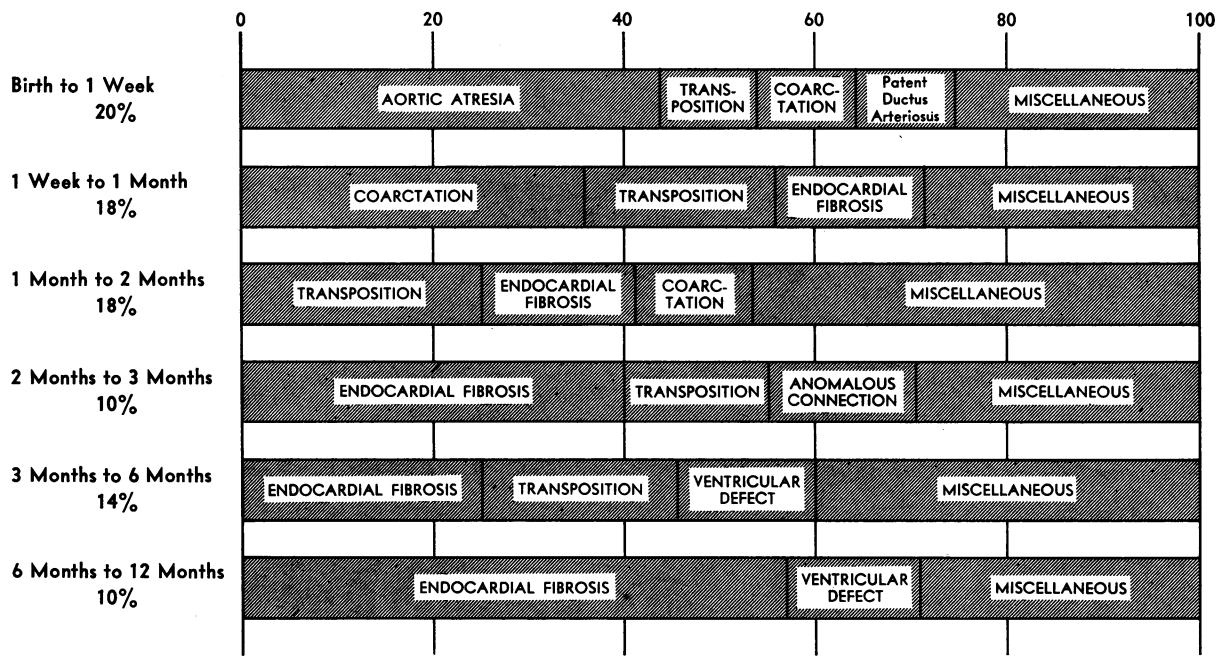

Chart 3.—Cause of heart failure correlated with age (304 cases). (Adapted from Keith: Pediatrics, 18:491, 1956.)

TABLE 5.—*Cause of Death Under 4 Years of Age (1,145 Cases);* No Operation Available at Present*

Diagnosis	Cases	Age at Death Less Than			
		30 Days	150 Days	1 Year	4 Years
Hypoplasia of left ventricle	63	43	60	63	
Truncus communis	48	33	42	47	1
Hypoplasia of aorta	42	21	36	39	3
Diverse	73	22	47	68	5
Total	226	119	185	217	9

*Adapted from Boesen and Vendel: VIII International Pediatric Congress.

failure in patients under one year of age. In addition to the high incidence of transposition of the great vessels and coarctation of the aorta as a cause of heart failure, the high incidence of endocardial fibroelastosis, particularly after the first month of life, was noteworthy.

It is becoming increasingly evident that in the instance of at least two congenital heart lesions, patent ductus arteriosus and interventricular septal defect, there is a far-advanced clinical situation in which surgical treatment by present methods is probably contraindicated. In the severe form of "reverse" patent ductus arteriosus, in which the major portion of the flow through the ductus arteriosus is from pulmonary artery to aorta, the operative mortality is approximately 100 per cent, for reasons not well understood.[8] In contrast, in cases in which the major portion or all of the flow through the ductus arteriosus is from aorta to pulmonary artery, the operative mortality is below 1 per cent.[11] Recently evidence was presented to suggest that patients with a partial "reverse" ductus arteriosus operated upon at an early age can survive the operation even though there is some reverse flow.[3] The main problem is early recognition and definitive treatment.

Eisenmenger complex has long been recognized as a form of interventricular septal defect. The main aberration that distinguishes it from simple interventricular septal defect is a systolic pressure in the right ventricle which exceeds that in the left ventricle, causing right-to-left interventricular shunt.[5] Most of the patients with nearly balanced pressures in the right and left ventricles have extensive changes in the intima and media of the pulmonary arterial tree.[5] Evidence recently presented[2] showed that the operative mortality in this group of patients also approximates 100 per cent, whereas it is much lower for patients who do not have the elevated right ventricular pressures.[2] Clear-cut evidence is not available to show that right ventricular and pulmonary hypertensions progress with time. However, severe forms of the disease with pulmonary hypertension are known to exist in the infancy and early childhood period. It is entirely possible that the pathological changes present in the lungs of younger patients with the disease will not be so extensive or irreversible at this earlier age.

University of California at Los Angeles School of Medicine, Los Angeles 24.

REFERENCES

1. Abbott, M. E.: Atlas of Congenital Heart Disease, American Heart Association, New York, 1936.

2. Adams, P., Lucas, R. V., Ferguson, D. K., and Lillehei, C. W.: Significance of pulmonary vascular pathology in ventricular septal defect as determined by lung biopsy. Presented before 29th meeting, American Heart Association, Cincinnati, Ohio, 1956.

3. Anderson, R. C., Adams, P., Jr., and Varco, R. L.: Patent ductus arteriosus with reversal of flow, Pediatrics, 18:410, 1956.

4. Boesen, I., and Vendel, S.: Scientific Exhibit, VIII International Pediatric Congress, Copenhagen, Denmark, 1956.

5. Dammann, J. F., Jr., and Ferencz, C.: The significance of the pulmonary vascular bed in congenital heart disease, III. Defect between the ventricles or great vessels in which both increased pressure and blood may act upon the lungs and in which there is a common ejectile force, Am. Heart J., 52:210, 1956.

6. Gardiner, J. H., and Keith, J. D.: Prevalence of heart disease in Toronto children, Pediatrics, 7:713, 1951.

7. Keith, J.: Congestive heart failure, Pediatrics, 18:491, 1956.

8. Kjellberg, S. R., Mannheimer, E., Rudhe, U., and Jonsson, B.: Diagnosis of Congenital Heart Disease, Year Book Publishers, Inc., Chicago, 1955, p. 426.

9. Maronde, R. F.: Brain abscess and congenital heart disease, Ann. Int. Med., 33:602, 1950.

10. Richards, M. R., Merritt, K. K., Samuels, M. H., and Langmann, A. G.: Congenital malformations of the cardiovascular system in a series of 6,053 infants, Pediatrics, 15:12, 1955.

11. Scott, H. W.: Closure of the patent ductus by surgical-ligation technique, Surg., Gynec. & Obst., 90:91, 1950.